

Five-Year Follow-Up of Peripartum Cardiomyopathy

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Abstract

Abstract- Peripartum cardiomyopathy is a type of cardiomyopathy found in pregnancy and up to 5 months after delivery. There is no identifiable cause for myocardial dysfunction in these patients. In this study, we evaluated our patients for symptoms, signs, functional class, prognosis and complications.

Methods- Eighteen pregnant patients with myocardial dysfunction and diagnosis of peripartum cardiomyopathy were evaluated from Dec. 1998 to Dec. 2007. All patients had echocardiography follow up, and symptoms and signs of every patient were evaluated for 5 years.

Results- Mean age of patients was 34 ± 5 yrs, mean left ventricular ejection fraction (LVEF) according to echo was $27\pm 5\%$, and dyspnea was present in 100% of patients. Chest pain was present in 61.11%, arrhythmia in 50%, edema in 72.22% and hypertension in 27.77%. During follow up, there were 22.22% next pregnancies in 5 years. Mortality occurred in 16.66%, remission in 27.77%, partial resolution of symptoms and signs and improved LVEF in 22.22%, and no improvement in 33.33%. Vaginal delivery was performed in 77.77% and cesarean section in 22.22% of patients.

Conclusion- Peripartum cardiomyopathy is a lethal disease during pregnancy. We do not recommend allowing next pregnancies and cesarean section is lethal; vaginal delivery is the best method of parturition for these patients (*Iranian Heart Journal 2008; 9 (1): 14-17*).

Key words: peripartum cardiomyopathy ■ vaginal delivery ■ cesarean section ■ dilated cardiomyopathy

Peripartum cardiomyopathy is a type of dilated cardiomyopathy that is seen in the last phase of pregnancy.¹ The syndrome has been defined by the following four criteria: 1) the development of cardiac failure in the last month of pregnancy or within 5 months of delivery, 2) absence of an identifiable cause for the cardiac failure, 3) absence of recognizable heart disease prior to the last month of pregnancy and 4) left ventricular systolic dysfunction demonstrated by classical echocardiography criteria, such as depressed ejection fraction.¹⁻² However there are some reports that early evidence of the disease was seen in the second and third trimesters.⁴

The incidence of this disease is reported between 1 per 4000 to 15000 deliveries, and the incidence is higher in Africans. Peripartum cardiomyopathy can be seen at any age, but is more common in women older than 30 years.⁵

The disease is related more to the first and second pregnancies and has strong correlation with gestational hypertension, twin pregnancy, and the use of tocolytic therapy.^{5, 6} Common symptoms and signs of disease are shortness of breath, fatigability, chest pain, palpitation, unusual weight gain, peripheral edema, and arrhythmias.⁶⁻⁸

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show tachycardia, ST-T wave changes, arrhythmias, and conduction abnormality.⁹⁻¹¹

On echocardiography, there is enlargement of the four cardiac chambers, depressed left ventricular ejection fraction, some degree of mitral and tricuspid regurgitation, and pericardial effusion.¹¹⁻¹⁴

Methods

In this study we followed 18 patients with documented peripartum cardiomyopathy from Dec. 1998 to Dec. 2007. All patients with a history of new dyspnea during the second or third trimester of pregnancy had echocardiography performed with 2 operators and ejection fraction of patients was estimated. Patients with abnormal ejection fractions were followed up one month later for further evaluation. All patients had new onset dypnea and no history of cardiac disease. After delivery, all patients were closely followed by a cardiologist and echocardiography was done every 6 months. All of them have been followed up for 5 years. The medications in pregnancy for all of the patients included digoxin, diuretics and hydralazine. After delivery all of them received digoxin, diuretics and ACEI or angiotensin receptor blockers. The patients were visited by cardiologists every month before delivery, and every week in the last month of pregnancy. Patients with a history of function class 4 were admitted in prenatal ward in the last month of pregnancy. All patients had cardiologist consultation for follow up during delivery and were followed for 5 years.

Results

There were 18 pregnant women (Table I). Mean age of patients was 34±5 years old (range 29-39 yrs). Mean left ventricular ejection fraction was 27±5% (range 22 – 32%).

There was dyspnea in all patients, and FC II was seen in 12 (66.66%), FC III in 3 (16.66%), and FC IV in 3 patients (16.66%). Chest pain was seen in 11 patients (61.11%), edema in 13 (72%) and arrhythmia in 9 patients (50%). Vaginal delivery was performed in 14 patients (77.77%) and 4 patients had cesarean section (22.23%). Next pregnancy occurred in 4 patients (22.22%) after 2 years of the first crisis, and all of them had successful vaginal delivery. Two patients had no aggravation of symptoms, but 2 patients were symptomatic with worse functional class. In echocardiography follow-up of patients, 5 (27.77%) patients recovered and left ventricular ejection fraction returned to normal within 6 months after delivery. There was relative clinical and echocardiography improvement in 4 (22.2%) patients but there was no improvement in 6 (33.33%) patients. Two (11.11%) patients died within 12 days of delivery by cesarean section; all patients that had vaginal delivery survived. However, one (5.55%) patient died at 4 months after vaginal delivery. Thirteen (83.33%) patients are alive at five years follow-up. Medications were continued in 10 (55.55%) patients for life and discontinued in 5 patients who recovered and have normal ejection fraction.

Table I. Demographic and outcome characteristics (n=18)

Age (year)	34
LVEF (%)	27±5
Mortality	3(16.66%)
Cure	5(27.77%)
Better out come	4(22.22%)
Unchanged	6(33.33%)
Vaginal delivery	14(77.77%)
Cesarean delivery	4 (22.22%)
Dyspnea	18 (100%)
FC I	0
FC II	12 (66.66%)
FC III	3 (16.66%)
FC IV	3 (16.66%)
Chest pain	11(61.11%)
Arrhythmia	9 (50%)
Edema	13 (72.22%)
Next pregnancy	4 (22.22%)
Hypertension	5(27.77%)

Discussion

Peripartum cardiomyopathy is a lethal disease that has mortality and morbidity in pregnancy.^{15, 16} In this study we evaluated the symptoms, signs and ejection fraction of our patients that have peripartum cardiomyopathy during 5 years. According to Fatkin and collaborators a third of cases of peripartum cardiomyopathy are inherited but in our study we did not evidence inheritance.¹⁵ Felker and colleagues performed endomyocardial biopsy in his patients but we did not perform this.¹⁴ In the study of Feldman and McNamara half of their patients with cardiomyopathy revealed myocarditis, but in our study we had no biopsy to document myocarditis.¹⁶ Barbaro and associates found 2 cases of HIV infection, but in our study we did not find any correlation between HIV infection and this disease.¹⁷ Cunningham found two thirds of his patients had hypertensive heart disease, but in our study we found this in 5 patients (27.77%).¹⁸ In Framingham heart study, Lauer and colleagues found a correlation between cardiomyopathy in pregnancy and obesity, but in our study we did not find this correlation. Lampert and Mabie implicated prolonged beta-mimetic tocolysis with terbutalin as provoking cardiomyopathy, but in our study we found no history of beta mimetic use in our patients.¹⁹ In the study by Sheffield and Cunningham, dyspnea was seen in all of his patients and chest pain was the second common complaint of patients, but in our study chest pain was the third complaint of our patients and peripheral edema was the second complaint.²⁰ Hibbard and co-workers found mean LVEF of patient to be less than 45%, but in our study it was 27±5%.²¹ In the study of Carlson and co-workers, some degree of deep vein thrombosis (DVT) was seen, but in our study we found no DVT in our patients.²² According to Lampert's study, half of his patients were cured during 6 months post-delivery, but in our study, this figure was only 5 patients (27.77%). Witlin and associates studied 28 patients and found two-

thirds had chronic hypertension and their outcomes were dismal: 5 patients died, 3 patient underwent heart transplant and 18 patients had continued cardiac impairment for a long time. Our study was in contrast to this report.²³ Albanesi and DaSilva described ventricular dysfunction in 3 of 12 women in subsequent pregnancy, but in our study 2 of 4 pregnancies had good outcomes.²⁴

Conclusion

Peripartum cardiomyopathy is a lethal disease in pregnancy. We found no evidence for inheritance pattern in our patients. Next pregnancies are not recommended for these patients. Cure rate in our patients is very low during 5 years' follow up. The best method of delivery for these patients is vaginal delivery; cesarean section is dangerous for this group of patients.

References

1. Lang RM, Lampert MB, Poppas A, William B: Peripartal cardiomyopathy .In Elkayam U, Gleicher N (Eds): Cardiac problem in pregnancy .3rd .New York, Wiley –Liss.1998, pp87-100.
2. Pearson GD, Veillle JC, Rahimtoola S, Mcain NC: Peripartum cardiomyopathy: National Heart, Lung, Blood Institute and office of Rare Disease (National Institute of Health) Work shop recommendation and review.JAMA 283:1183, 2000.
3. Akhtar WM, Shotan A, Hameed A, Villis M,Cooper J: Pregnancy associated cardiomyopathy: Early versus late presentation Am Coll Cardiol 41: 1039, 2003.
4. Fett JD: Peripartum cardiomyopathy .Insights from Haiti regarding a disease of unknown etiology. Minn Med 85:46,2002
5. Akhtar WM, Shotan A, Hameed A, Villis M,Cooper J: Pregnancy associated cardiomyopathy: Clinical profile in 137 patient diagnosed in U, S .Jam Coll Cardiol 41; 1136, 2003.

6. Aziz TM, Burgess mi, Acladiious NN, Millton J: Heart transplantation for per partum cardiomyopathy: A report of three cases a literature review. *Cardiovasc Surg* 7: 565, 1999.
7. Shotan A, Widerhorn J, Hurst A, Elkayam U: Risk of angiotensin-converting enzyme inhibition during pregnancy: Experimental and clinical evidence, potential mechanism, and recommendation for use. *Am J Med* 96: 451, 1994.
8. Ford RF, Barton JR, O Brien JM, Morgan M: Demographics, management, and outcome of peripartum cardiomyopathy in a community hospital. *Am J Obstet Gynecol* 182: 1036, 2000.
9. Carlson KM, Browning JE, Eggleston MK, O BrienJM: Peripartum cardiomyopathy presenting as lower extremity arterial thromboembolism: a case report. *J Report Med* 45: 351, 2000.
10. Bozkurt B, Villaneuva FS, Halubkov R, Millan N: Intravenous immune globin in the therapy of peripartum cardiomyopathy. *J Am Cpll Cardiol* 34: 177, 1999.
11. Sliwa K, Skudicky D, Candy G, Villis MN, Morgan M: the addition of pentoxifylline to conventional therapy improve outcome in patient with peripartum cardiomyopathy. *Eur J Heart Fail* 4: 305, 2002.
12. Sliwa K, Skudicky D, Bergemann A, And Thomson RE: Peripartum Cardiomyopathy: A nalysis of clinical outcome. left ventricle function, plasma level of cytokines and Fas /APO-O. *J Am Coll Cardiol* 35: 701, 2000
13. Felker GM, Thompson RE, Hare JM, Hamilton MN, Millkan GM: Underlying cause and long term survival in patient with initially unexplained cardiomyopathy. *N Engl J Med* 342: 1077, 2000.
14. ElkayamU, Tummala PP, Rao K, Morison JO, Morgan MN: Maternal and fetal outcomes of subsequent pregnancies in women with peripartum cardiomyopathy. *N Eng l J Med* 344: 1567, 2001.
15. Fatkin D, Mac RaeC, SasakiT, Wolff MR, Porcu M, Frenneaux M, Atherton H, Vidaillet HJ, Spudiich S, de Girolami U, Seidman JG, Seidman CE: Missense mutations in the rod domain of the lamin a/c gene as cause of dilated cardiomyopathy and conduction system disease. *N Engl J Med* 341: 1715, 1999.
16. FeldmanAM, Mc Namara D: Myocarditis, *N Engl J Med* 343: 1388, 2000.
17. Barbaro G, Di Lorenzo G, Grisorio B, Barbarini G :Incidence of dilated cardiomyopathy and detection of HIV in myocardial cells of HIV-positive patient. *N Engl J Med* 339: 1093, 1998.
18. Cunningham FG, Prichard JA, Hankins GDV, Anderson PL: Idiopathic cardiomyopathy or compounding cardiovascular event. *Obstet Gynecolo* 67: 157, 1986.
19. LampertMB, Hibbard J, Weinert L, Briller J: Peripartum heart failure associated with prolonged tocolytic therapy. *Am J Obstet Gynecol* 168: 493, 1993.
20. Sheffield JS, Cunningham FG: Diagnosing and management peripartum cardiomyopathy. *Contemo Ob Gyn* 44: 74, 1999.
21. Hibbard JU, Lindheimer M, Lang RM: A modified definition for peripartum cardiomyopathy. *Obstet Gynecol* 94: 311, 1999.
22. Carlson KM, Browning JE, Eggleston MK: Peripartum cardiomyopathy presenting as lower extremity arterial thromboembolism. *J Report Med* 45: 351, 2000.
23. Witlin AG, Mabie WC, Sibai BM: Peripartum cardiomyopathy: A m J Obstet Gynecol 176: 182, 1997.
24. Albanesi FM, Da Silva TT: Natural course of subsequent pregnancy after peripartum cardiomyopathy: *Arq Bras Cardiol* 73: 47. 1999.